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of the

CHILDREN'S HOSPITAL

WASHINGTON, D. C.

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VOLUME VIII NUMBER 2

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CLINICAL PROCEEDINGS

OF THE CHILDRENS HOSPITAL

13th and W Streets, Washington 9, D. C.

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THE TUMOR BOARD AND TUMOR CLINIC AT THE CHILDREN'S HOSPITAL, WASHINGTON, D. C.

David F. Bell, Jr., M.D.

The organization of the Tumor Board and Tumor Clinic in 1949 at the Children's Hospital has made possible a more standardized approach to the problem of malignant disease in childhood with resultant benefit to the patient.

The Tumor Board offers the advantage of scheduled (Wednesday, 2:15 p.m.) consideration of cases suspected of having malignant disease. Because cancer often runs a more rapid course in younger children than in the older age groups, the Tumor Board also has made it possible to obtain emergency consultation. Therefore, delay in diagnosis and therapy is unwarranted in cases suspected of having a malignant tumor. The Board has helped to crystallize methods of treatment of the various types of malignant disease. The chemotherapeutic, steroid, and other types of treatment of leukemia and malignant disease have been carried out under the supervision of the Board and the Department of Pathology.

It is not the purpose of the individual tumor board members to "force" their recommendations upon private or resident physicians referring patients to the Tumor Board. The Board acts in an advisory capacity only and feels that their considered opinion may be of value to the patient, the patient's family, and to the referring physician.

We have been fortunate at this hospital to have, in active participation on the Tumor Board, physicians with experience in pediatric pathology, radiology, and surgery. Tumors in children present problems necessitating decisions by physicians specializing within the field of pediatrics.

Through the activities of the Tumor Board, there has been brought to the community and the staff of the hospital, an increased awareness of the importance of malignant tumors and leukemia in children. This need for increased interest in tumors in children has been furthered from the educational standpoint by extending an invitation to all residents and medical students in the hospital to attend the weekly Tumor Board meetings. Any physician is welcome to attend. Exhibits have been prepared by the Tumor Board members from information obtained from the records of our patients with malignant disease and displayed at the annual Scientific Assembly of the Medical Society of the District of Columbia. In 1950, the Exhibit consisted of a graphically illustrated case report on one of our leukemia patients. Our display in 1951 gave the results of an extensive research (from the year 1938 to the present) on all patients with Wilms' tumors and neuroblastomas, the most common malignant intra-abdominal tumors in childhood.

Other advantages have become apparent following the organization of the Tumor Board and Tumor Clinic. Funds for the diagnosis, treatment, and care of children with tumors are more readily available to institutions with programs aimed at the diagnosis and treatment of malignancies. The American Cancer Society and the Damon Runyon Cancer Fund both have made funds available to The Children's Hospital for the support of the Tumor Board and Tumor Clinic and for the care of patients with tumors.

The Tumor Board and Tumor Clinic at this hospital have been approved by the American College of Surgeons with the recommendation that all tumors be registered with the Tumor Clinic, particularly in reference to private cases. The members of the Tumor Board and those active in the Tumor Clinic are anxious to comply with this recommendation and they would like to take this opportunity to ask for the registration of all tumor patients with the Tumor Clinic or Tumor Board, if only for statistical purposes.

The Tumor Clinic serves as an adjunct to the Tumor Board and it also meets weekly (Friday, 10:00 a.m.). Patients referred to the Clinic are examined with special reference to apparent or suspected tumors. Such patients are screened—given a complete physical examination and workup before presentation to the Tumor Board. Approximately 80 per cent of the cases seen in the Clinic are subsequently presented to the Board. Complete records are kept on file and much effort is devoted to the continuous follow-up of diagnosed cases. Particular care is taken in following patients with malignant tumors and these have a follow-up record of about 90 per cent. A name card file is kept on all patients as well as a diagnostic card file, coded in accordance with *The Standard Nomenclature of Disease*. A full-time secretary is employed by the Tumor Board for the purpose of carrying out this work.

The problems of diagnosis and treatment of malignant disease in children are important because of the changing causes of death in childhood. Infectious diseases have always been associated with children, and before 1947 more deaths occurred from the infectious and parasitic diseases than from cardiac diseases and cancer. For a number of years there has been a decreasing mortality from the infectious diseases and since 1947

YEAR	TOTAL NUMBER OF PATIENTS SEEN		PATIENTS WITH MALIGNANT DISEASE	
	Tumor Board	Tumor Clinic	Tumor Board	Tumor Clinic
1949	51	69	31	31
1950	84	83	24	17

cancer and cardiovascular diseases have accounted for more deaths than the infectious and parasitic diseases combined [1].

REFERENCE

1. Public Health Reports, Vol. 66, No. 39 (September 28) 1951.

ADRENOGENITAL SYNDROME

Case Report No. 224

Edwin C. Shepherd, M.D.

The purpose of this paper is threefold: 1. To stress mixed adrenal disease as it occurs in infancy by reporting another case. 2. Further, to indicate that even in the presence of fairly typical signs and symptoms, the diagnosis of this disease may be overlooked until too late. 3. To reemphasize that the syndrome is one that, when diagnosed early, can be treated specifically with resultant preservation of life, as evidenced by the increasing accumulation of favorable reports in the literature.

CASE REPORT

An eighteen-day old white male patient entered the hospital because of weight loss. The history revealed that the child was born spontaneously without instrumentation or difficulty after approximately seven and one-half to eight months' gestation. The birth weight was 5 pounds and the color and cry were good. Delivery took place in the family doctor's clinic, and the mother and child were sent home the next day. After three days at home the child was returned to the clinic because the mother thought he was having abdominal pain. The patient remained there four days before being sent home on a 1:4 evaporated milk formula. The mother stated the baby ate well, taking 12 ounces of the above formula each day. At eighteen days of age the patient was taken back to the family doctor for a routine check-up, at which time it was discovered that he weighed 4½ pounds, and had not eaten well that morning. It was also noted that he had a slight discharge from both eyes which had been present for three or four days. The patient was then sent to this hospital for admission.

Physical examination on admission revealed no remarkable findings other than a poorly developed and moderately malnourished eighteen-day old white male patient weighing 4 pounds, 2 ounces with good color and strong cry. The skin was warm and dry; turgor was fair, but the skin folds were loose, and there was a minimum amount of subcutaneous fat. There was no rash. There was a moderate amount of purulent discharge from both eyes, and the sclerae were mildly jaundiced. The edge of the liver was palpable. The genitalia were that of a male with incomplete fusion of the scrotal sac at the anterior midline just below the base of the penis. The testes were not felt either in the sac or inguinal canals.

The admission diagnoses were: 1. prematurity, 2. moderate malnutrition, and 3. moderate conjunctivitis.

The patient was placed in an incubator with continuous oxygen and was started on glucose water by mouth until a formula of 1:2 olac could be prepared. He was

then offered 2 ounces of formula every three hours, the amount to be increased as tolerated. Aqueous procaine penicillin, 200,000 units daily was also started. Smears and cultures were taken from the eyes, but no organisms were found. The eyes were washed three times daily with a solution of boric acid water.

The admission hemogram revealed 13.8 grams of hemoglobin; 5,000,000 red blood cells; 14,800 white blood cells with 47 segmented cells, 6 band forms, and 47 lymphocytes per 100 cells. The platelets appeared normal. The red blood cells showed anisocytosis.

The patient did well throughout the first and second hospital days, taking his formula with increasing eagerness and he seemed to become more and more active with the passage of time. Hydration seemed satisfactory. During the early hours of the third hospital day, the patient became somewhat listless. This progressed through the morning hours until he became markedly listless and acquired an ashen color. He took very small amounts of formula and vomited most of this. Respirations became irregular and shallow with periods of apnea, and slight cyanosis became evident. Dehydration seemed marked. He was started on gavage feedings and was given intravenous and subcutaneous fluids. In the afternoon he had improved considerably, and on the morning of the fourth hospital day he seemed in fairly good condition, taking over 2 ounces of formula without gavage. Respirations had become regular and normal; the heart and lungs were clear. Supplemental parenteral fluids were maintained, and on the fifth hospital day the patient's condition was thought to be satisfactory. During the early morning hours of the sixth hospital day the patient again began to vomit, did not eat well, became progressively more listless and apathetic, became dehydrated and appeared to be in poor condition. More parenteral fluids were given and the hemogram was repeated, this latter showing no remarkable change other than that of hemoconcentration. Respirations again became shallow with periods of apnea, cyanosis appeared, and the patient died shortly thereafter.

With the onset of the second episode of rapid dehydration and shock-like state and in the presence of malformation of the genitalia, the diagnosis of adrenogenital syndrome was given strong consideration, and with resultant death, this diagnosis was felt to be the most tenable one. Unfortunately, it was made too late. At autopsy, the pathologic diagnosis was: 1. pseudohermaphroditism secondary to 2. adrenal hyperplasia with proliferation of the androgenic zone.

DISCUSSION

Deficiency disease of the adrenal glands has been recognized for nearly a century, the classical signs of pan-hypoadrenalism having been described by Thomas Addison in 1855.[1] The work of Albright,[2] Kendall,[3] Selye,[3] Thorn,[4] and others [23, 24] indicates that various functions of the adrenal cortex exist and, as a result, a number of types of adrenal insufficiency have been revealed, not all of which show the complete picture of Addison's disease. Total obliteration of adrenal function is thought to be rare in childhood and, when occurring, is extremely acute and usually fatal. In the Waterhouse-Friderichsen syndrome, complete or partial destruction with hypo-function of the adrenals occurs and death often results. Since described by Butler,[5] Ross and Talbot,[5] Wilkins, Fleshmann and Howard,[6] and Thelander and Cholffin,[7] mixed

adrenal disease is receiving increased attention and many cases, both from the clinical and pathologic point of view, have now been reported in the literature.[7–18] The usual picture is one in which there is hypofunction or absence of the mineralocorticoids, increased androgenic corticoids, and probably normal glucocorticoids. However, it has been postulated [19, 20] that deficiency states might exist including the glucocorticoids. A case of glucocorticoid insufficiency has been described in infancy by Geppert, Spencer, and Richmond.[21]

The etiology of mixed adrenal disease is not clearly understood, although several theories have been advanced. The suggestion, that the persistence of the fetal or androgenic zone into postnatal life might be responsible for the excess adrenal androgens, remains unsupported by most investigators.[5, 7, 13, 18, 22] Knudson,[23] in a recent generic study,

TABLE 1

Symptomatology

USUAL SYMPTOMS	SYMPTOMS PECULIAR TO FEMALES	SYMPTOMS PECULIAR TO MALES	FUTURE EXPECTATIONS IF LIFE MAINTAINED
Vomiting Diarrhea Dehydration Acidosis Visible peristalsis Weight loss Circulatory collapse Cyanosis	Pseudo-hermaphroditic changes	Sexual precocity (after a few months)	Increased rate of somatic growth; precocious puberty; premature closure of epiphyses; ultimate short stature; pigmentation[5, 9, 13, 26, 23] (though rarely)

discloses good evidence that the disease is inherited recessively and he developed the concept of an inborn error of adrenal steroid metabolism.

It is not uncommon for the onset of symptoms to occur in the first few days or weeks after birth. Also, it is important to emphasize that in some cases these symptoms have led to the incorrect diagnosis of pyloric stenosis, intestinal obstruction, or some vague parenteral infection. For this reason, it is imperative that the practitioner always keep the syndrome keenly in his awareness. See Table 1.

Laboratory findings, where obtained, usually show an increase in the urinary 17 ketosteroids in both sexes. The serum electrolyte determinations reveal lowered sodium and chloride values, while the potassium values are increased.[13, 26, 27, 30] These values may be normal except during crisis.[23] The blood sugar, though sometimes lowered, is usually not significantly so. Serum bicarbonate levels are reduced. Urinary chlorides are increased.

Mixed adrenal disease of infancy is often misdiagnosed or not suspected.

It must be considered in an infant who has persistent vomiting for no apparent reason or in which some of the symptoms already described are evident. It then becomes necessary to determine certain laboratory data, for it is by this means, keeping in mind the clinical picture as it may present itself, that the diagnosis can be more accurately pinpointed, and the more common diseases simulating this syndrome, e.g., pyloric stenosis, primary cardiac disease, renal disease, temporary adrenal insufficiency as described by Jaudon,[28] or other rarer adrenal insufficiency states can be excluded.

To preserve life, it is urgent that treatment be started as soon as possible. Parenteral saline and sodium lactate correct the electrolyte loss. Desoxycortico-sterone acetate is felt to be most effective in preventing renal electrolyte loss. It is believed that large doses are necessary, 2.0 mgms. daily being the average.[23] However, Mullings and Chute[19] have used adrenal cortical extract, later switching to desoxycortico-sterone acetate in oil with success. They have also had gratifying results with the use of desoxycortico-sterone acetate pellets implanted subcutaneously. Therapy for female virulism or male precocity in association with accelerated growth remains generally unsatisfactory, though adrenalectomy[11] and preliminary cortisone therapy[21] have both been tried.

SUMMARY

1. A case of mixed adrenal disease clinically suspected but not diagnosed until autopsy has been presented.

2. A discussion of the review of the literature has been made.

3. It is emphasized that this syndrome, if kept in the mind of the practitioner, can be diagnosed and will frequently respond to treatment, which is specific.

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TYPHOID FEVER IN SIBLINGS SUCCESSFULLY TREATED WITH CHLOROMYCETIN

Charles L. Waite, M.D.*
David L. Simon, M.D. L. R. 51-10827
John P. McGovern, M.D. V. R. 51-10896

The purpose of this report is to point out several facets in the response of typhoid fever to a relatively new antibiotic, to reemphasize the natural history of this disease as modified by age in childhood and to demonstrate what can be accomplished in tracing the epidemiology of this disease

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through the cooperation of the county and out-of-state health departments.

L. R., an eight year old white male, was admitted to the Children's Hospital on September 17, 1951 with the chief complaint of unremitting fever of two weeks' duration. The onset of illness was gradual with malaise, headaches, anorexia, and temperatures ranging from 101.0 F. to 104.0 F. Six days before admission, treatment was started by his private physician with penicillin and terramycin in unknown amounts, but the latter was discontinued after one day because of a diarrhea that developed. A blood culture was taken three days before admission; therapy with penicillin was continued. When it became apparent that the child was not responding to therapy, penicillin was discontinued and he was admitted to the hospital.

Physical examination at the time of admission revealed a well developed and well nourished child appearing acutely ill. The temperature was 104.0 F. rectally. There was marked lassitude yet the remainder of the physical examination was within normal limits. The red blood cell count was 4,600,000 with 12 grams of hemoglobin. The white blood cell count was 9,100 with a normal differential. Cerebrospinal fluid taken on admission was normal. The blood culture taken three days before admission revealed Salmonella typhosa. Febrile agglutinations taken on admission were positive for typhoid "O" antigen in a dilution of 1:1280 and for the "H" antigen in a dilution of 1:160. Chloromycetin therapy was started on the day of admission in the dosage form of 250 milligrams immediately and 250 milligrams every six hours. The fever continued as did the malaise, anorexia, and severe headaches. On the third hospital day, the chloromycetin was increased to 500 milligrams every six hours. A stool culture taken on the same day grew out S. typhosa. Approximately thirty-six hours after increasing the dosage, the temperature returned to normal and remained normal for the rest of the hospital stay. The patient received chloromycetin until the twenty-second hospital day. The only complication that developed during the course of the illness was a secondary anemia (2,800,000 red blood cells and 9.2 grams of hemoglobin). Supportive therapy consisted of polyvitamins and he received a blood transfusion on the twenty-fifth hospital day. He was discharged after twenty-six days of hospitalization.

On September 19, 1951, V. R., a six-year old sister, was admitted to this hospital with a chief complaint of fever and headache of two weeks' duration. Her temperature had previously been recorded as high as 105.0 F., but at the time of admission had dropped to 98.4 F. rectally. On the day before admission it had risen to 104.0 F. Physical examination revealed a slightly enlarged spleen. The remainder of the physical examination was within normal limits. Repeated blood and stool cultures for S. typhosa were negative, but the agglutination for typhoid "O" antigen was positive in a dilution of 1:1280. The patient received an average of 1.5 grams of chloromycetin per day for nineteen days. Her course in the hospital was uneventful, the temperature remaining normal throughout the hospital stay, and she was discharged on the twentieth hospital day.

These two siblings live in Silver Spring, Maryland, a suburb of Washington, and had the advantages of modern sanitation offered this community. When the positive agglutinations were reported, the Montgomery County Health Department was notified and a systematic search for possible sources of the infection was begun. A small stream closeby was tested and eliminated as the source. Daily stool cultures were taken on the members of the immediate family and were found to be negative. Two brothers, both in the Armed Forces, were examined and were found to be free

of infection. Finally, the county health department, working through the Iowa State Health Department, received notification that positive stool cultures had been obtained from the grandfather living in Des Moines. He had visited the family in Silver Spring three weeks prior to the onset of the illnesses, and is currently being treated as a carrier.

DISCUSSION

Prior to the use of Chloromycetin, there was no successful specific therapy for typhoid fever. Even though the disease has decreased in incidence in the United States in the last twenty-five years, due to improved sanitary and immunological measures, the mortality rate has remained unchanged. It is approximately 5 per cent in children and young adults and 10 per cent in infants one year and under. It is too early to say exactly to what degree the mortality rate will be affected by the use of Chloromycetin. However, from a general review of the literature of the past three years, it is becoming increasingly evident that death from typhoid fever which has been treated early and to a sufficient degree will be considerably lessened. Though it seems apparent that Chloromycetin acts specifically in controlling this bacterial infection, it must be remembered that it is not highly bactericidal toward S. typhosa and that it acts primarily as an inhibitor. It should also be kept in mind that this mode of therapy does not hasten the healing of intestinal ulceration and that relapses and persistently positive stool cultures will occur although no more frequently than in the natural course of the disease.[1, 2]

In a series of 17 cases of typhoid fever treated with Chloromycetin at this hospital since 1949,[3] several interesting features in the clinical response not previously reported have been noted. First, there is a definite delay in the initial response of the patient who is being adequately treated with Chloromycetin. This delay or "lag-phase" usually lasts 62 hours or slightly longer, during which time the patient continues to have a spiking temperature and other evidences of active disease. It is felt that many of the so-called "therapeutic failures" that are reported were due to failure to recognize this peculiarity in response This resulted in the early discontinuance of the Chloromycetin before its efficacy could be noted. At present there has been no investigation recorded concerning the possible reasons for this "lag-phase" following the institution of therapy. The generally accepted dosage form is 100 milligrams per kilogram of body weight each twenty-four hours. Failure to continue the treatment for an adequate length of time may result in a recurrence. Treatment with adequate levels of Chloromycetin should be continued for five days following three negative stool cultures. Many recurrences may actually be due to reinfection, and, in order to prevent this every effort should be made to trace the source of infection.

The clinical manifestations of typhoid fever in children are, for the

most part, similar to those in the adult, but usually are less severe. The wide variation in the incubation period (three to forty days), often makes the task of tracing the source a difficult one. Fever and malaise are the most predominant symptoms in the child, and diarrhea is present in 50 per cent of the cases. In the series studied at this hospital diarrhea was the exception rather than the rule. Constipation is frequently one of the chief complaints. A palpable spleen and rose spots aid in making the clinical diagnosis but are not always detectable. Infection in the neonatal period may present a severe sepsis or diarrhea alone, but because the opportunity for contact is so limited these cases represent less than 1 per cent of those occurring in the first year of life.

The diagnosis depends upon blood, urine, and stool cultures, and after the tenth day of illness, upon the Widal agglutination test. A Widal test should be run on every case at the time of admission, not only because of the long incubation period, of the disease, but also to establish a base line for the "H" and "O" antigens, a rising titer of the "O" antigen being a most significant feature in making the laboratory diagnosis.

Complications are fortunately less frequent in children than in adults. The most common complication noted at this hospital in the series treated with Chloromycetin was a hypochromic microcytic anemia of the type seen following many infectious diseases in children. There were no cases of intestinal hemorrhage or perforation of the bowel. Delirium and coma are not uncommon. Exanthemata, alopecia, and grooving of the nails are occasionally seen.

The two cases reported here serve to reemphasize an oft-neglected aspect in the overall treatment of this disease, namely, that of tracing down the source of the infection. Typhoid fever occurring in one or more members of a family with a relatively high standard of living in a community where modern methods of sanitation are practiced should make the physician suspicious of a carrier as the likely source of infection. In this instance, the stream near the house, around which the children frequently played, was found to be free of pollution. Two brothers, both in the Armed Forces were found to be free of infection, but the grandfather who had visited the house three weeks before the onset of the illness was found to be a carrier. Such a result in determining the source of the infection in this case is a tribute to the excellent and integrated public health setup of the various federal, state, county, and municipal agencies of the United States.

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IMPERFORATE ANUS AND ASSOCIATED MALFORMATIONS

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Anorectal malformations are an important group of anomalies responsible for many deaths during the first few days of life. There are two embryologic abnormalities responsible for the condition: failure of the cloacal duct to close, resulting in a connection between the rectum and the genito-urinary tract, or persistence of the anal membrane, causing imperforation of the anus.

Estimates regarding the frequency of these conditions vary between 1 in every 4,000 to 10,000 live births.[1, 4, 5] Associated anomalies often occur; even multiple anomalies are not uncommon. In some series, these were present in as many as 68 per cent of the cases,[5] and were directly responsible for some of the deaths. Congenital heart disease and atresia of the esophagus or elsewhere in the intestinal tract are the most common offenders.

Anorectal anomalies can be divided into four types.

TYPE I

The anus is patent but incomplete obstruction exists. The stenosis occurs at the mucocutaneous junction or lower rectum and is due to incomplete rupture of the anal membrane. There is no actual atresia. Patients with this condition come under observation later than those of the other types unless the stenosis is so marked that it produced intestinal obstruction.

Fig. 1a

Diagnosis: Obstipation with varying degrees of abdominal distension and a ribbon-like character to the stools are the usual findings. The perineum may bulge on straining if the stenosis is marked. The point of narrowing can always be felt with the examining finger.

Treatment:

1. Repeated dilatations—daily at first—gradually decreasing in frequency.
2. Incision longitudinally with transverse closure of the point of stenosis.
3. Colostomy is limited to the very few patients in whom the above

methods are unsuccessful, or whose general condition does not permit more definitive surgery.

4. If a fistula does exist, the rectum should be mobilized downward and the fistula exteriorized.
5. Rarely is a combined abdomino-perineal "pull-through" procedure necessary.

Results in these cases are good. They have normal sphincter muscles. The reported mortality is less than 10 per cent.

TYPE II

FIG. 1b

There is an imperforate anus but the obstruction is membranous. It is due to a persistence of the anal membrane. This is the least common type of anomaly and in an analysis of 214 cases, only 9 had this type of malformation.[1]

Diagnosis: Establishment of the diagnosis is not difficult since there is no anal opening and the baby manifests classical signs of intestinal obstruction. No stools are passed. The abdomen becomes distended and vigorous peristalsis and vomiting are noted. Marked bulging of the perineum occurs on straining and often a dark discoloration in the region of the skin dimple can be seen. This is caused by the pooling of meconium in the lower end of the blind rectal pouch.

Treatment:

1. Incision: cruciate through the skin dimple.
2. Excision of the membrane and suture of the mucosa to the skin edges.
3. Dilatations may be used if steps 1 and 2 are not entirely successful.

Results: This group of patients are the simplest to treat and the overall results are good. The mortality is approximately 10 per cent and all of the patients have normal sphincter muscles.

TYPE III

The anus is imperforate, but its site is marked by a skin dimple. The rectal pouch ends blindly a variable distance above the perineum. This is the most common type of anomaly and approximately three-fourths of all patients fall into this category. It is this type of case in which most of the fistulas occur. The symptoms are those of complete intestinal obstruction unless a large fistula is present.

FIG. 1c

Physical Examination: The anal site usually consists

of a puckering or skin dimple. If puckering is present, the existence of sphincter muscles is established. The presence or absence of intestinal obstruction depends on the existence and size of an associated fistula. In some of these cases, especially with rectovaginal or rectoperineal fistulas, the position of the blind pouch may be determined by passing a probe into the fistula or by introducing barium into the tract under x-ray examination. An impulse on straining may be felt in the perineum if the pouch is low.

Treatment depends on the location of the rectal pouch, the presence or absence of a fistula and the infant's general condition. Primary perineal repair is the treatment of choice. It consists of a perineal approach with downward mobilization of the rectum which is sutured to the skin edges after the sphincter muscles have been reconstructed about it. To be successful, the blind end of the rectum must not be separated from the perineum by too great a distance. It is essential that there be no tension on the suture line. Ladd and Gross[1] consider this critical distance to be 3 centimeters while Koop[2] and others[5] feel that 1½ to 2 centimeters is the maximum distance which can be successfully bridged without fear of retraction.

Results are often poor in these cases. Sphincter control may be absent. Dense scarring due to retraction of the bowel up into the pelvis often results in secondary obstruction. The mortality is about 25 to 30 per cent.

TYPE IV

The anus and lower rectum are normal. The upper rectum ends blindly a variable distance above the lower rectal pouch. Detection is difficult because the anus appears normal. Surgical intervention often is delayed several days, therefore, these cases represent poor surgical risks.

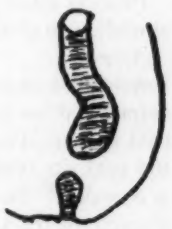

FIG. 1d

Physical Examination: The upper limit of the lower rectal pouch can always be felt on digital examination. Sphincter muscles are present and normal. Intestinal obstruction with distension, vomiting, and dehydration is common.

Treatment: These cases are difficult to treat because of the patient's poor condition. The following measures have proven to be the most successful in the correction of this anomaly:

1. Colostomy.
2. End-to-end anastomosis.
   A. perineal approach
   B. abdominal approach
3. Abdomino-perineal "pull-through" operation.

Results: The reported mortality in this group of patients is over 50 per cent. Sphincter control is present in most cases.

DIAGNOSIS

The diagnosis is established by inspection, palpation, and x-ray examinations. The perineum should be examined for discoloration, dimpling, puckering of the skin, or bulging. Low fistulas (rectoperineal or rectovaginal) are usually easily identified while the existence of a rectourinary fistula may be difficult to demonstrate. If a rectourinary fistula is suspected, gross and microscopic examination of the urine will aid in establishing the diagnosis.

Most of these infants come under observation because of the absence of an anus or the presence of an abnormal one; the failure to pass meconium; or the passage of meconium through an abnormal opening. Absence of an anal opening may be overlooked for days and the situation may be discovered only when an attempt is made to introduce a thermometer or to give an enema. If a large fistula exists, the absence of an anal orifice may escape detection for years.

Symptoms: At birth, almost three-fourths of these patients have actual or impending complete intestinal obstruction. These attract attention early and are classed either as type I with extreme stenosis or as any of the other three types with an absent or very narrow fistula. The remaining patients are seen later and are grouped as type I with minimal stenosis or as any of the other types with large fistulas.

Physical Examination: Varies in each case and is included in a discussion of each type of anomaly.

X-rays:

A. Air contrast studies—The position of the blind rectal pouch may be determined by taking an x-ray of the abdomen and pelvis with the baby held feet up. Air rises in the bowel and with the aid of a metal marker at the anal site, the distance between the terminal bowel and the perineum is revealed.[10] To avoid errors in diagnosis at least eighteen hours must be allowed for the air to reach the lower limit of the bowel.

B. Barium studies—The status and position of the rectal pouch may be demonstrated by x-rays taken after barium has been injected through the fistulous tract or into the distal colon if a colostomy has been performed.

ASSOCIATED ANOMALIES

Associated anomalies often occur in this condition and even multiple anomalies are not uncommon. The associated anomaly may be directly responsible for the infant's death. The ten most commonly encountered

associated anomalies are:

1. Congenital heart disease
2. Esophageal atresia
3. Hypospadius
4. Atresia of the small bowel
5. Malformed vertebra
6. Other penile anomalies
7. Septate vagina
8. Absent metatarsal bones
9. Hare lip and cleft palate
10. Meckel's diverticulum.

FISTULAS

Associated fistulas between the rectum and the genito-urinary system or the perineum are fairly common. In large series of patients with im-

I. Rectovesicle

II. Rectourethral

III. Rectoperineal

Fistulas in males
FIG. 2.

I. Rectovesicle

II. Rectovaginal

III. Rectoperineal

Fistulas in females
FIG. 3.

perforate anus, about 50 per cent are found to have fistulas. They occur more frequently in females than in males. Most of the fistulas are in the type III anomaly; e.g., in one review of 117 cases, all except 6 fell within

N.B. FIGURES 1, 2, and 3 redrawn from Ladd and Gross: Abdominal Surgery of Infancy and Childhood, W. B. Saunders Co., Philadelphia, Pa., 1941.

this category.[1] Although fistulas may be present in all four types, they are encountered most infrequently in the type IV malformation. Multiple fistulas are rare but they do occur.

The size of the fistula varies. Usually, it is so small that partial or complete intestinal obstruction exists, necessitating operation during the first few days of life. However, it may be so large that it goes undetected for years.

In the female, most of the fistulas are rectovaginal, located in the lower half of the vaginal or in the interlabial region. Rectourinary fistulas, on the other hand, are rare in females, but quite common in males. They may escape detection. A narrow tract may close temporarily or, if the bowel obstruction is relieved, it may close permanently. Rectoperineal fistulas occur with about the same frequency in both sexes.

Rectourinary fistulas should be closed as soon as possible to avoid urinary tract infections. Colostomy will divert the fecal stream and some small tracts will close if the imperforate anus is corrected. Combined abdominoperineal repair allows correction of both conditions at one operation. Rectovaginal and rectoperineal fistulas are usually closed at the initial operation.

TREATMENT

Several accepted methods of management are available in treating an infant with one of these anomalies. The method one selects depends on the general condition of the patient, the type of anomaly, and whether or not a fistula is present. If a method requiring more than one operation is selected, timing of these procedures is of the utmost importance. The operative risk during the first seventy-two hours of life is not great, but after this time, mortality increases dramatically.

The management differs somewhat in the two sexes, only because in the male, the problem usually is one of complete intestinal obstruction and immediate operation is mandatory. In the female the presence of a rectovaginal fistula may allow one to delay operation indefinitely. Dilatation of small fistulas permits procrastination, but Koop and others feel that this delay should not be more than three months. By this time, the fistula should be closed and the anus repaired. Operation can be delayed longer but complications such as urinary tract infection and dilatation of the colon can be expected—to say nothing of the increasing anxiety of the parents. Rectourinary fistulas rarely are large enough to relieve the intestinal obstruction.

Some of the methods of treatment are:

1. *Dilatation.* When the anomaly is a type I anal stenosis, repeated dilatations will usually suffice. If a low fistula (rectoperineal or recto-

vaginal) is present, definitive surgery may be indefinitely delayed by stretching the fistula to allow emptying of the bowel.

2. *Primary perineal repair.* All workers agree that a primary perineal repair is the treatment of choice if it can be done with ease and safety. In determining whether or not a primary perineal repair can be accomplished, the distance separating perineum and rectal pouch must be measured. Ladd and Gross feel that any distance greater than 3 centimeters is too great to be successfully bridged from below, while Koop considers this critical distance to be 1½ centimeters. It is important to utilize an incision which will cause minimal damage to the sphincter muscles and yet will allow adequate exposure. Dissection must be carried high into the pelvis to allow mobilization of the bowel down to the perineum without tension. If this is not accomplished, the retaining sutures will invariably cut through; the terminal rectum will retract, and a scar will form. A longitudinal incision extending from the forchette back to the skin dimple or even to the coccyx is the customary one. Resuturing of the divided sphincter muscles is essential.

3. *Colostomy.* Primary colostomy is usually limited to infants weighing less than 5 pounds, those in poor condition, or those with high fistulas. Secondary colostomy is resorted to after attempted perineal repair has failed. With the aid of antibiotics, colostomy now is not dangerous. In spite of this, it is being employed with less frequency than in the past because the trend is away from "staged" procedures. If a colostomy must be performed, it is essential that the descending or sigmoid colon be spared for later use. The left half of the tranverse colon is the site of election. A terminal colostomy rather than a loop or tangential colostomy is imperative for complete diversion of the fecal stream

4. *Combined abdominoperineal repair.* A one stage "pull-through" operation is safe and effective and now is being employed frequently. Its place is in the treatment of patients in whom the blind rectal pouch is too high to allow primary perineal repair, or in those cases with high associated fistulas. The use of this procedure avoids multiple operations. Rectourinary fistulas may be closed at the same time the imperforate anus is repaired.

Mortality. Review of the literature [1, 8, 9, 2, 5] reveals the mortality in the various surgical procedures to be as follows:

Primary successful perineal repair	12–22 per cent
Primary colostomy	18–25 per cent
Secondary colostomy following unsuccessful perineal repair	68–85 per cent
Combined abdominoperineal repair	10 per cent

Case Report K. L. D. 51-8388

K. L. D. was the product of a spontaneous, full term delivery. At birth she appeared normal except for an imperforate anus with a rectovaginal fistula. A skin

FIG. 4. Preoperative photograph showing imperforate anus with skin dimple. The vagina is normal and the orifice of the fistula is visible.

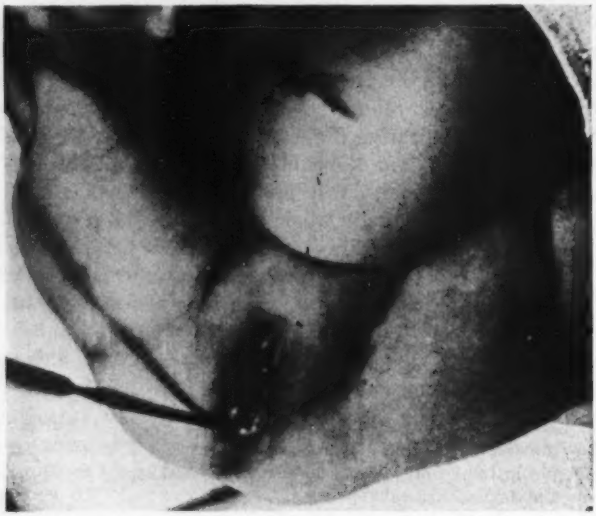

FIG. 5. Preoperative photograph showing a metal probe introduced into the opening of the fistula.

dimple with obvious sphincter muscles was present, but there was no discoloration or bulging of the perineum on crying. Several hours after birth she began passing meconium through the low rectovaginal fistula. For three weeks she continued to have bowel movements which were ribbon-like in character. She then became obstructed.

First Admission. On July 18, 1951 she was admitted to Children's Hospital, Washington, D. C. because of abdominal distension and failure to have bowel move-

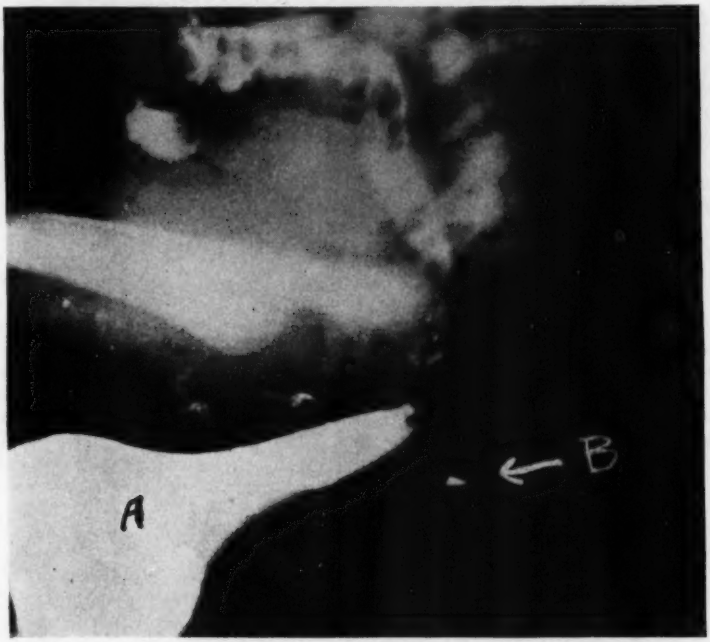

FIG. 6. Preoperative x-ray showing the outline of fistulous tract and colon with barium. The distance between the skin marker and the rectal pouch is 3 centimeters. A. Bulb syringe with barium. B. Metal marker on skin dimple. C. Fistulous tract. D. Terminal rectum.

ments for eighteen hours. Physical examination revealed a healthy, well developed and well nourished three week old infant. The abdomen was slightly distended; there was an imperforate anus; a skin dimple and sphincter muscles were present. A small opening with a tract leading back towards the rectum could be seen in the region of the fossa navicularis between the labia and just outside the hymen. A soft rubber catheter was introducted through the fistulous tract and a warm water enema was given. This was followed by the expulsion of a large amount of semi-solid fecal matter. The distension was relieved and the child's condition improved. Diagnostic studies were begun the next day. A thin barium mixture was introduced through the fistulous opening and the lower rectum was examined by means of

fluoroscopy and x-ray. The fistulous tract was shown to make a straight upward course from the lower vagina up to the blind rectal pouch. A metal marker on the perineal skin dimple revealed the rectum to end 3 centimeters above the perineum. This was classified as a type III anomaly with an associated low rectovaginal fistula. It was decided to dilate the fistulous tract temporarily and to attempt a primary perineal repair within a few weeks. This dilatation was easily accomplished by

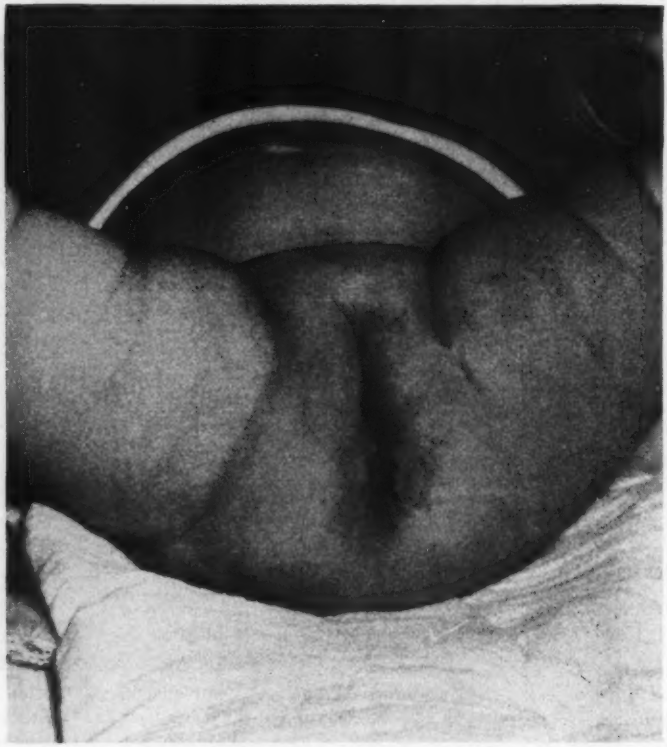

FIG. 7. Postoperative photograph revealing the repaired anus; the fistula is closed. A small amount of rectal mucosa is visible.

introducing a curved clamp into the tract and gently stretching the point of narrowing. The child was then discharged from the hospital to be followed at home. At this time she was having normal spontaneous stools through the fistula.

Second Admission. On September 18, 1951, the patient was readmitted to the hospital. At this time she was three months old; her progress had been satisfactory, and physical examination was essentially the same as on the first admission. She was prepared for operation with enemas and intramuscular chloromycetin and penicillin.

Operation: September 19, 1951. The patient was anesthetized with venethene,

oxygen, and ether and placed in the lithotomy position. A longitudinal perineal incision was made extending from the forchette back to the center of the anal skin dimple. Dissection was carried down to the external and internal sphincter muscles which were divided in the anterior midline and retracted laterally and posteriorly. Great care was exercised to preserve these muscles intact posteriorly. Using blunt and sharp dissection, the fistulous tract was identified, dissected free, and amputated at its entry into the vagina. The vaginal fistulous stoma was closed with two layers of catgut sutures. Then, aided by downward traction on the fistulous tract, the rectum was mobilized up through the pelvis almost to the peritoneal reflection. It was densely adherent to the upper posterior vaginal wall, but was entirely free elsewhere. Upon completion of the dissection, the rectum and proximal portion of the fistula were mobilized downward, the distal portion of the fistula was cut away. A perineal body was constructed; the sphincter muscles, reunited in the anterior midline. The submucosa of the bowel was attached to the sphincter muscles with several interrupted sutures. The mucosa was sutured to the skin edges, and the perineum was loosely closed without drainage.

Postoperatively, the patient was fed parenterally and given nothing by mouth for twenty-four hours. On the second postoperative day she was started on liquids by mouth. About six hours after operation she began having bowel movements per rectum. Intramuscular chloromycetin and penicillin were continued for six days. The postoperative course was entirely uneventful and she was discharged from the hospital on September 27, 1951—eight days after the perineal repair of her imperforate anus with closure of the rectovaginal fistula.

Gentle, rectal dilations were begun on the fourteenth postoperative day and have been continued daily until the present time. These are performed at the same time each day by the mother who inserts her index finger into the rectum leaving it in place for about five minutes. The child has gained weight, has eaten satisfactorily, and the bowel movements have been normal. The rectal mucosa and skin edges have healed together nicely. A small amount of mucosa is visible when the child is placed in the lithotomy position. The rectal tone appears normal.

SUMMARY

1. The various types of anorectal anomalies are discussed.
2. The types and management of associated fistulas are outlined.
3. The diagnosis and treatment of these conditions are presented.
4. A representative case of imperforate anus with rectovaginal fistula, successfully repaired, is included.

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